UPPER DYSPHAGIA*

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THE œsophagus has been termed, not inaptly, the step-child of practical medicine and, like its prototype, it has suffered unduly from neglect. Looking upon it as clinically unimportant, the average practitioner still finds it hard to take active interest in it. This perhaps applies more particularly to its upper opening where it is joined by the hypopharynx. Text-books and periodical literature give it little notice and mainly focus attention on the lower segments, and much has been written upon so-called cardio-spasm. A well-known continental authority, in an address two or three years ago upon what a progressive practitioner ought to know of œsophageal affections, passed over the upper orifice merely with a reference to cancer and pressure pouch. This attitude can no longer be justified; increasing knowledge has removed much of the mystery that shrouded the organ. New methods of examination have enlarged our horizon and it might not be inappropriate to attempt a short synthesis of the stages leading up to this beneficial advance and discuss briefly some of the results achieved. It may stimulate students of the problems involved, to whom the wider issues have perhaps grown dim, to consider afresh some points of contact that have been linked up with more than one branch of medicine. My early training has impressed me with its value. There is much to be said for the historical approach in medicine, so let me recall some of the steps of our enlightenment.

The examination of the upper food passage has always been dependent upon mechanical aids, the use of which goes back to early times. Tongue depressors and spatulae were necessarily limited in range and real advance came with the laryngeal mirror. The evolution of the œsophagoscope, as it is to-day, we owe to the untiring efforts of the Vienna school, and especially to Stoerk and von Mikulicz. Of the problems

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concerned, whilst that of light was solved by technical advances in illumination generally, the means of its conduction downwards into the closed œsophagus went through various phases before the simple rigid straight tube was finally adopted. Stoerk quite early interested himself in the question and constructed both flexible and rigid tubes at different times and continued to use both forms during his life-time for diagnostic purposes and extraction of foreign bodies. He made repeated efforts to devise a practical flexible tube to avoid the discomfort of the rigid one which he likened to " the barbarous method of sword-swallowing ". He was a pioneer of the method but as he published little he failed to establish its general use.

The real introduction came in 1881 by von Mikulicz, a general surgeon, interested in the surgery of the stomach and œsophagus. Familiar with Stoerk's work, and using straight tubes with improved illumination, he showed the value of the method and, by demonstration in 1882, definitely established its practicability. Von Hacker, who followed him, removed a large number of foreign bodies and taught the feasibility of inspecting the trachea and bronchi by passing the tube through the larynx. In 1894, Kirstein, working with a tube-spatula and improved lighting, achieved a direct view of the larynx and devised an autoscope which I found practicable only when pressure on the tongue could be tolerated. It remained for Killian in 1897 to pick a foreign body out of the right bronchus and establish the value of bronchoscopy and, with it, endoscopy of the upper food and air passages was placed on a sound footing.

Radiography has made rapid advances in œsophageal work in recent years. The ordinary method of filling out the canal with barium paste, by which we recognize stenosis, dilatation, and pouching, is no longer sufficient and, for delineation of the mucosa, relief representation, extended to the œsophagus by Wolff in 1928 and developed later by Berg, Schatzki and Palugay, is indispensable. By using barium in small quantity and of suitable consistency, folds and irregularities of the mucous surface, varices and small ulcers, etc., are outlined and it is an excellent way of locating non-opaque foreign bodies, such as chicken bones and fish bones, that elude detection by ordinary means. The administration of a small dose of atropine, by drying the mucosa and slowing down movement, reduces the speed of passage through the upper orifice and

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permits more careful observation. Schatzki's recently published studies[1] on the hypopharynx and mouth of the œsophagus, are testimony to its value. The air-relief representation of Palugay,[2] a combination of the relief method with presence of air in the canal is another striking development. Both methods give informative pictures and, though not without difficulties in carrying out, are a welcome addition to our means of diagnosis.

The new methods, by making possible collation of the results of observation in the living with those in the cadaver, have contributed to our knowledge of the parts about the upper orifice. Killian, usually so accurate a writer, is incorrect in thinking that the mouth of the œsophagus, marked by the stellate-shaped contracted lumen is closed by the lower portion of the inferior constrictor of the pharynx. Elze,[3] holding the view that such sphincters are, with few exceptions, not morphological but functional formations, gives reasons for believing that the true functional constrictor producing the characteristic stellate lumen as seen through the endoscope, is the upper circular fibres of the œsophagus. Further, he regards as a misnomer the term " lip of the œsophagus ", applied to the sickle-shaped fold on the posterior hypopharyngeal wall obstructing the view through the œsophagus and offering difficulties to its introduction. It can take no part in the mouth as it lies 2-3 cm. above it. " Hypopharyngeal fold ", he thinks, would more accurately describe what is really a large submucous plexus which only becomes apparent in the cadaver after the parts are injected and is therefore usually overlooked.

Turning to questions awaiting solution I propose to limit my remarks to certain conditions of the hypopharynx and upper opening of the gullet which produce dysphagia. One of the most interesting, and perhaps the least recognized in adult life, is congenital narrowing of the œsophagus. From the clinical standpoint the subject is not without difficulty. We are dependent upon the history given by the patient of long-standing impairment of swallowing. Slight inability may have existed from early life, necessitating a restriction of food to which the patient has become accustomed, and regards as normal. Not infrequently attention is first drawn to it by some mischance such as food sticking in the passage. I recorded,[4] a good many years ago, the case of a lad, 17 years

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of age, having a history of a " small swallow " for years and who was brought to hospital totally unable to swallow after eating meat. On removal of a piece of beef impacted behind the cricoid plate, a narrowing of the gullet immediately below could be seen. After the local disturbance had subsided, difficulty with solids remained as before and it seemed more than probable that we were dealing with a congenital condition.

A number of cases of congenital narrowing without history of dysphagia have been recorded in the literature. Brown-Kelly[5] discussed the subject five years ago at the Royal Society of Medicine, and showed a *post mortem* specimen from a man of 35 who had never had trouble in swallowing and had served at the front in the War, yet the lumen at the pharyngo-œsophageal junction had a diameter of only 4 millimetres. The remarkable instance reported by D. M. Greig[6] of a woman of 82, who had never been able to take solid food and lived only on liquids and whose death was due to senility, showed that good development and absence of emaciation were compatible with a narrowing of the œsophagus at the cricoid level, found after death, through which a small probe could be passed with difficulty. The case illustrates an adaptation to the restricted passage of food that may remain through life, an inconvenience causing no symptoms so long as the diet is fluid and accidents are avoided. In such individuals due appreciation of the congenital limitation and suitable modification of diet throughout life form the basis of treatment. It is of practical importance to remember that well-masticated solid food can be passed through a lumen of 5 mm. in diameter according to Jackson, who, writing recently,[7] further says that a malignant stricture of 3 mm. lumen will take almost all food ordinarily eaten by the average man provided it has been put through a sieve of 25 meshes to the inch.

The problems raised by the foregoing lead to consideration of a much larger group, also having dysphagia as a cardinal symptom, where organic disease is not usually thought to be in evidence. The comprehensive term " functional dysphagia " of the textbooks comprises a large undifferentiated group embracing types ranging from hysterical vagaries to more persistent inability to swallow. It is common experience that cancer of the œsophagus is not infrequently diagnosed at some stage as hysteria. A great step forward in our

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knowledge came with the introduction of the new modes of examination and, in 1906, in a paper[8] on the direct method, then in its infancy, I referred to a type of dysphagia of " so-called functional character, occurring chiefly in women and associated often with atrophic catarrh of the upper pharynx ", in which a definite diagnosis was possible only after direct examination, and quoted an illustrative case. It soon became possible to distinguish as a separate entity a clinical form of dysphagia, frequently looked upon hitherto as functional, which was subsequently described by Brown-Kelly[9] and myself[10] in papers published in 1919, the main features of which I may be permitted to recall.

It affects women of middle age, not regarded as hysterical, in whom a long history of slowly increasing inability to swallow, often associated with anæmia and atrophic changes in the mucosa of the mouth, tongue and upper pharynx are prominent symptoms. The pallor of the patient, sometimes profound, the mouth with cracked lips and fissures at the angles giving it a contracted look, the tongue changes that range in individual cases from mere smoothness to advanced glossitis combine to produce a characteristic picture.

The local appearances, viewed through the œsophagoscope, are by no means uniform. The loss of the normal sphincter-like form of the mouth of the œsophagus with its soft mucous folds is not unusual and the entrance appears as a pinhole or irregular opening or slit or partly closed by a thin mucous web or tense band readily ruptured by passage of the endoscope. Or there may be a feeling of resistance to the tube suggestive of spasm. Such variability may be due partly to the pressure of the tube, partly to an altered mucosa. To the latter change can be ascribed a want of resiliency not infrequently present that may be correlated with a thickening of the submucosa observed microscopically by me in the buccal and lingual mucous membrane. In most cases, passage of a tube into the œsophagus is followed by improvement in swallowing and there is an improvement in the patient's condition that is often striking.

The incidence of this symptom-group—dysphagia, atrophic change and anæmia—from the standpoint of the laryngologist may be gathered from figures taken from my hospital out-patient notes in twelve consecutive years from 1908 to 1920, omitting 1910 in which the notes are incomplete. The

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number of patients in their respective age-groups is as follows :—

DYSPHAGIA.

Age-Group.	*Number of Patients.*
20-30	7
30-40	18
40-50	28
50-60	12
60-70	1
	66

Of the 66 patients over two-thirds of the number fall into the age-group 30-50, the youngest being 22, the oldest 62. The average duration of symptoms is difficult to estimate. A considerable number of patients gave a history of " a small swallow " all her life or for many years. The shortest period is over a twelvemonth. Some complained of inability amounting to little more than inconvenience until aggravated by an accident due, in six cases, to impaction of meat. Dysphagia in the youngest patient, a married woman of 22, came on at the age of 19 after the birth of the first child. Co-existent affections such as heart disease, asthma, floating kidney and uterine trouble are noted in a few.

Nearly all presented atrophic change in mouth and pharynx as well as anæmia, but, and this must be emphasized, in varying degree. In some, one or both symptoms were scarcely noticeable. Direct examination revealed the usual diversity in the picture and, when made a second or third time after some months' interval, the appearances did not always coincide. In several instances a further inspection, after lapse of a year or more, revealed the supervention of malignant disease.

In a paper in 1914 Plummer gave the name of hysterical dysphagia to a group of cases characterized by a sense of obstruction at or about the œsophageal entrance, nearly always associated with pallor and secondary anæmia and frequently enlargement of the spleen. Under the same title, Vinson[11] in 1922 reported a series of sixty-three patients suffering from dysphagia, three-fourths being males and one-fourth females in whom moderate secondary anæmia was present. He noted nothing abnormal by endoscopy in the upper food passage and no mention is made of atrophic change

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in the mouth or of glossitis. The condition was regarded as hysterical, treatment consisting in the passing of a bougie which was used purely as a psychological measure and the administration of iron and arsenic.

In 1926 Dr. A. F. Hurst published a case which, as he remarks in a paper in an American journal[12] two years ago, was described by him " under the rather unfortunate name of the Plummer-Vinson syndrome—unfortunate, because I subsequently discovered that excellent accounts of the condition, though without any details as to the type of anæmia present, had been published in 1919 by Brown-Kelly and Paterson."

The type of anæmia associated with this affection was first differentiated in 1913 from the group of secondary anæmias, and became known in this country by the excellent work of Witts[13] and Dan Davies,[14] the former suggesting the name simple achlorhydria anæmia which has passed into general use. Affecting women between the ages of 30 and 50 and comparatively common, it is of the hypochromic microcytic type, characterized by marked reduction of the hæmoglobin content, though the red cells are little below normal, and accompanied, though not invariably, by achlorhydria. When marked the spleen may be palpable. The sallow appearance, fissures at the angles of the mouth, and atrophic tongue mucosa are regarded as valuable diagnostic features, and brittleness of the nails with a tendency to split and become spoon-shaped are said to be characteristic. Even with a minor degree of anæmia, glossitis is rarely absent. In 101 cases reported by Witts, dysphagia was present in thirteen. The anæmia is said to be associated with chronic gastritis, and the clinical features suggest a nutritional disorder, possibly an iron deficiency.

The dysphagia is, in all probability, related to change in the mucosa though this may not always be well-marked. Brown-Kelly believed stenosis to be due to a firm approximation of folds and not to organized adhesion. Hurst, on the other hand, cited a want of relaxation of the sphincter—achalasia—as the cause. A loss of resilience in the parts, by impeding free movement of the mucosa on the underlying loose submucosa, is no doubt a contributory factor to which may be added the possibility of muscular paresis comparable to the atony that can be induced artificially by atropine. In this connection the striking effect of the passage of a tube

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in relieving obstruction and materially improving the patient's condition may be recalled.

The question of the order in which the symptoms in the syndrome appear has provoked a good deal of discussion. In the earlier papers, both laryngologists and physicians agreed in regarding dysphagia as the initial factor. The patient not unnaturally stressed it and thought of it as the cause of her poor health. Later, however, with the differentiation of hypochromic anæmia and recognition in recent years of its frequency in women, physicians came to view it as primary. The history given by the patient, which is generally thought to settle priority, is not always convincing and it is not unusual to witness in these cases a vicious circle in which anæmia and dysphagia react on each other and form a feature of the illness.

The distribution of the affection calls for some notice. Until three years ago, when contributions first began to appear in continental literature, its bibliography had been confined to English and American sources and there was a tendency to regard it as an Anglo-Saxon disorder. Neither physicians nor laryngologists in Germany appear to have differentiated it and Henning,[15] in a paper in 1935, on œsophageal affections refers to it as little known. Nor is it listed in French or Italian literature. On the other hand, important papers appeared in 1933 and 1935 by Wassink of Amsterdam who approached it from a different angle and a contribution on similar lines has come this year from Stockholm.

This brings me to another aspect—the relation of the syndrome to malignant disease. It is now recognized that malignant disease shows a striking tendency in women to involve the hypopharynx and mouth of the œsophagus and this termination is no unusual event in the affection under consideration—a point to which attention was called by Kelly and myself in our original papers. Logan Turner's[16] statistics established the greater incidence of malignant disease at the upper end of the œsophagus amongst women and at the lower end in men, that in post-cricoid carcinoma 86 per cent. of the cases are women and that the highest age-incidence falls between 30 and 50 in women and between 50 and 70 in men. My own figures, though much smaller in number, taken from hospital notes over twelve consecutive years, confirm this.

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Pirquet[17] in his admirable analysis of English statistics lends support to these conclusions. He points out that of deaths from œsophageal cancer in the period 1911-26—24 per cent. were in women—men are in greater proportion in the middle cancer-period and women in the earlier period. Their graphs also are different, those of men rising steeply to a peak and those of women showing a plateau or double curve summarizing two different disposition-periods, the younger age with an upper localization in the œsophagus and the higher age with localization lower down.

Post-cricoid cancer occurs less frequently in German and French statistics. This may be due to some extent to difference in nomenclature and it is noteworthy that in few continental countries has the sex incidence of œsophageal cancer been worked out. Differences in population groups no doubt account for much. According to a statistical law of cancer, holding good for all civilized societies with reliable statistics, while the organ incidence may show wide differences the total incidence is nearly the same for all. This variability in the organ incidence explains much of the variation in different populations, but its underlying cause eludes the most detailed and exhaustive inquiry.

Wassink[18] has thrown fresh light upon the question from another aspect. As surgical director of Leeuwenhuis, the Cancer Institute of Amsterdam, he made careful investigation into the early history of cases of cancer of the hypopharynx and mouth of the œsophagus in women and obtained evidence of early difficulty in swallowing long before the disease became manifest. He described types, closely reproducing the main features of the syndrome. The small capacity for swallowing for years, the typical facies, atrophic changes, etc., are exact parallels. It is improbable, in view of their early appearance, that they were the result of malignant disease. He does not regard dysphagia as the cause but the indirect consequence of the anæmia.

Further support has recently come from the large material of Radiumhemmet, the Cancer Institute of Stockholm, where investigation on similar lines made by Ahlbom[19] confirmed the existence of a relationship between the syndrome and squamous carcinoma of the upper end of the food tract.

In a communication presented to the recent International Cancer Congress in Brussels, Wassink remarks on *le fait*

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curieux, as he terms it, that in the literature relating to the syndrome of dysphagia with anæmia and the distinctiveness of cancer of the hypopharynx in women, with the exception of some Swedish observers and himself, one finds only the observations of Anglo-Saxon writers. Would this, he asks, point to an actual rarity of these cases in the other countries and ought one to see in it an expression of an idiosyncrasy peculiar to the peoples mentioned, who are certainly linked demographically.

It remains to emphasize the value of treatment. This calls for close co-operation between physician and laryngologist. The anæmia should be dealt with by the administration of iron and it is an axiom that it be given regularly, by the mouth, and in large doses—60 to 90 grains of iron and ammonium citrate over a long period. Small doses are of little use. The condition of the tongue may be improved by liver extract. Reference has been made to the good effect of passing an œsophageal tube or bougie in relieving the dysphagia and this may have to be repeated. The importance of maintaining nutrition by food, suitable in form and quantity, has already been stressed.

Recently the problem of relieving these cases has been approached by Lambert Rogers[20] from another side. After pointing out that the blood supply of the anæmic atrophic mucosa of the hypopharynx and the œsophageal orifice, as well as the innervation of the sphincter, are under the control of the sympathetic component of the pharyngeal plexus derived entirely from the superior cervical ganglion, he suggested that the removal of the ganglion on both sides would result in increase of the blood supply and relaxation of the sphincter. This he carried out in a well-marked case which had been subjected to the usual treatment with little effect and the patient states that she has now lost the feeling of dryness in the mouth and pharynx and is able to swallow better.

To sum up, we may define this type of dysphagia as primarily related to change in the mucosa the nature of which is not quite clear. As laryngologists we are familiar with the far-reaching results of prolonged dysphagia. On the other hand, its association with a secondary anæmia, of obscure origin, in women of reproductive age, and often accompanied by atrophic alteration in mouth, pharynx and nails giving the patient a

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characteristic appearance, seems to suggest a wider basis and an underlying, possibly constitutional, factor. I have sufficiently stressed the not infrequent supervention in these cases of malignant disease in the post-cricoid region with its distinctive features of a much higher incidence and earlier age in women and the question has been raised whether an effective antecedent treatment of the anæmia by iron would not prevent this distressing sequel. These and other intrinsic problems, touching more than one branch of practical medicine, are too important to be allowed to remain a mere catalogue and it is within the province of laryngology to make contribution to their solution.

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Die hier besprochenen Schlingbeschwerden sind solche, die auf krankhaften Schleimhautveränderungen beruhen. Diese Veränderungen stehen in Zusammenhang mit einer sekundären Blutarmut unklaren Ursprunges bei noch gebärfähigen Frauen bei denen oft auch Schwunderscheinungen im Mund, Schlund und an

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den Nägeln vorkommen, die ihnen ein eigenartiges Aussehen verleihen und den Gedanken nahelegen, dass es sich um eine angeborene Anlage handelt.

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Le type de dysphagie qui est discuté dans cet article est celui qui est en relation avec des modifications de la muqueuse. Elle est associée à de l'anémie secondaire d'origine obscure, chez des femmes à l'âge de la reproduction, et elle est souvent accompagnée d'une modification atrophique de la bouche, du pharynx et des ongles, donnant aux malades un aspect caractéristique, ce qui fait soupçonner l'intervention d'un facteur constitutionnel.

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